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Unique Presentation of Submandibular Gland Adenoid Cystic Carcinoma: Case Report and Review of Literature

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Abstract:-

- ***Background;***

Adenoid Cystic Carcinoma (ACC) is a rare tumor, accounting for less than 1% of head and neck malignancies and approximately 10% of all salivary gland tumors. Following the parotid and sublingual glands, the submandibular gland is the most commonly affected site (Spiro et al., 1974). These tumors are frequently devoid of symptoms, exhibit slow growth, and are known for their characteristic infiltrative growth pattern and perineural invasion..

- ***Case Detail;***

This research presents a case of Adenoid Cystic Carcinoma in a 45-year-old female who presented with a left submandibular swelling persisting for 12 years. The diagnosis was confirmed through histopathological examination, and the study also includes a review of the distinctive clinical and histopathological characteristics found in the literature.

- ***Conclusion***

Salivary gland tumors can manifest in any location where salivary tissue is present. Adenoid cystic carcinoma is a rare type of salivary gland tumor known for its varied histomorphological characteristics. Achieving an early diagnosis and developing a treatment strategy requires thorough patient history assessment, clinical examination, and the incorporation of radiographic and histopathological findings(Spiro, 1986; Spiro et al., 1974).

Keywords:- *Adenoid Cystic Carcinoma, Salivary Gland Cancer, Radiation Therapy, Local Recurrence.*

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I. INTRODUCTION

Adenoid cystic carcinoma is a malignant neoplasm that can affect both major and minor salivary glands in the oral cavity. In 1853 and 1854, three Frenchmen named Robin, Lorain, and Laboulbene independently documented this condition (Coca-Pelaz et al., 2015). They were the first to observe the cylindrical appearance of this tumor. In 1859, Billroth described ACC as "cylindroma" due to its cribriform appearance, characterized by tumor cells forming cylindrical pseudolumina. He also noted its propensity for recurrence. ACC accounts for approximately 4% to 11% of all salivary gland neoplasms and represents 3% to 5% of malignant head and neck tumors. Minor salivary glands, primarily in the palate but also in the submandibular and parotid glands, are affected in about 30% of cases. Although it can occur across a wide age range, it predominantly affects women in their 5th and 6th decades of life (Vikram et al., n.d.).

Typical clinical manifestations include slow growth, local recurrence, perineural invasion, and distant metastasis. ACC presents in three histological subtypes: cribriform, tubular, and solid. These subtypes may occur independently or concurrently within the same tumor, with the solid subtype being the most aggressive. Tumors are graded based on the criteria proposed by Szanto et al. as cribriform or tubular (grade I), less than 30% solid (grade II), or greater than 30% solid (grade III). ACC is widely recognized as a highly challenging disease to manage, with Conley referring to it as "one of the most biologically destructive and unpredictable tumors of the head and neck." In this report, we present a case of adenoid cystic carcinoma of the submandibular gland and provide a brief review of the clinical, histopathological, and therapeutic aspects of this condition.(Cai et al., 2014; Spiro, 1986)

II. CASE REPORT

A 45-year-old woman sought medical attention at the oral and maxillofacial surgery department of Rama Hospital, presenting with a primary concern of a left submandibular swelling that had persisted for 12 years. She reported no difficulties with chewing or swallowing. The swelling had started as a small, painless lump and had gradually grown in size over the course of 12 years. An intraoral examination revealed no abnormalities, and her mouth opening was normal. There was no evidence of any surface discharge. Upon palpation, the swelling exhibited characteristics of being painless, firm in texture, and movable. Routine blood tests, including CBC, PT, INR, LFT, and KFT, returned within normal parameters. A preliminary diagnosis of a submandibular gland tumor was made. Additionally, occlusal and panoramic radiographs did not reveal any alterations in bone structure. USG Guided FNAC Suggestive of Salivary gland neoplasm, MILAN Category IV-B, i.e., SUMP (salivary gland neoplasm of undetermined malignant potential). On CT face with contrast, there was evidence of a large lobulated heterogeneously enhancing soft tissue density lesion in left submandibular space with multiple non enhancing areas within it. It measures approximately 46 x 32 x 40 mm in size (APTRCC). Large lobulated heterogenously enhancing soft tissue density lesion in left submandibular space. Findings are suggestive of infective etiology more likely over neoplastic etiology. Few homogenously enhancing subcentimeter sized nodes are noted in la, bilateral level Ib, I1. Resection of tumour was done under General anaesthesia and was sent for histopathological examination. On microscopic examination it shows a diffusely invasive tumour disposed in cribriform pattern with focal tubular and sheet like pattern of neoblastic basal cells/myoepithelial cells. Cribriform spaces are filled with eosinophilic hyaline material. Foci of perineural invasion and lympho-vascular tumour emboli are seen. Stroma show abundant Myxo-hyaline material. Adenoid Cystic Carcinoma (cribriform pattern) was the final diagnosis.

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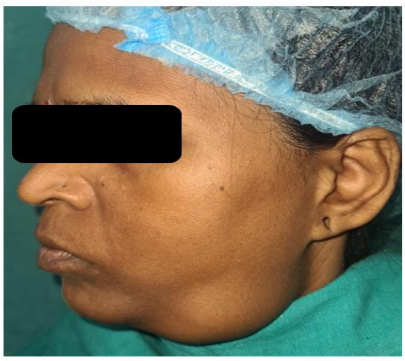

Figure: A
Swelling in submandibular region

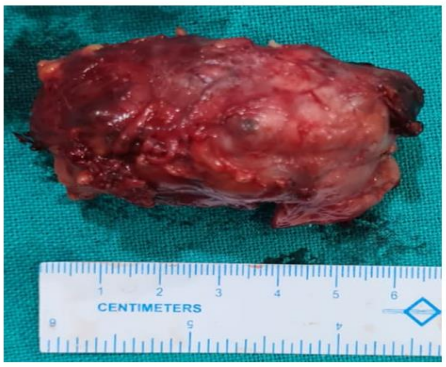

Figure: B
Resected tumor

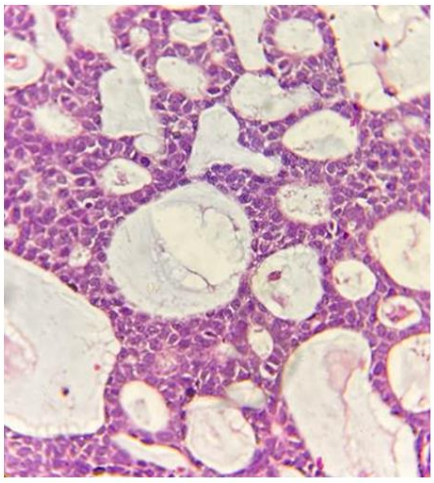

Figure: C
The histopathological analysis of the mass in the right submandibular gland reveals a nodular growth of uniform basaloid cells arranged in a cribriform pattern, which is indicative of adenoid cystic carcinoma.

III. DISCUSSION

Adenoid cystic carcinoma (ACC) represents a rare subtype of adenocarcinoma, accounting for approximately 31.6% of all minor salivary gland tumors(Rafael et al., 2016). The most commonly affected location is the palate, with the tongue being the second most frequent site. ACC predominantly arises in minor salivary glands, with a higher incidence in the submandibular gland and less frequently in the sublingual and parotid glands. Intraosseous tumors in the maxilla and mandible are exceptionally rare occurrences(Kumar Subudhi et al., n.d.). ACC is believed to originate from glands that secrete mucus and involves cells capable of differentiating into both epithelial and myoepithelial cells. Structures originating from the foregut, specifically the parotid, submandibular, and sublingual glands, as well as mucus-producing glands in the upper respiratory tract, are exclusive sites for mucus-secreting tumors(Choi et al., 2022; Rodriguez-Russo et al., 2021). Typically, these tumors measure around 2-4 cm at their largest dimension, with intraoral adenoid cystic carcinoma rarely exceeding 3 cm. The lesions can present as non-encapsulated and infiltrative, often involving bone invasion(Coca-Pelaz et al., 2015). The likelihood of cervical metastasis is relatively low, while distant metastasis typically occurs via the bloodstream, affecting the lungs and bones. In some cases, direct extension of the lesion into the base of the skull has been identified as a cause of fatality(Choi et al., 2022). There are three histopathological patterns: cribriform, tubular, and solid. The cribriform pattern is the most prevalent, characterized by epithelial cells arranged in multiple cylindrical spaces. The tubular type consists of ducts formed by one or two layers of cells resembling myoepithelial cells. The solid variant comprises solid epithelial islands with central areas of necrosis. Cells in the solid variant are small, basophilic, hyperchromatic, with densely granulated nuclei and few mitotic figures. Prognostically, the cribriform type generally carries a favorable outlook, whereas the solid variant is associated with the poorest prognosis(Kumar Subudhi et al., n.d.).

The primary treatment approach is extensive surgical removal, with the addition of radiotherapy if lymph node metastasis occurs(Vikram et al., 1984). Key prognostic factors include the size of the primary lesion (T), its anatomical location, the presence or absence of metastasis (M) at the time of diagnosis, invasion of the facial nerve, and the histopathological grade (G). Treatment options encompass surgical therapy, radiotherapy, chemotherapy, and combinations thereof (such as surgery and radiotherapy or radiotherapy and chemotherapy).(Vikram et al., 1984) Relying solely on surgical excision or radiotherapy may result in potential relapse within surgical margins and the development of metastases in cervical lymph nodes, lungs, or bones. Adenoid cystic carcinoma also exhibits a tendency to invade nerves near the lesion due to its neurotropic characteristics. Salivary gland neoplasms represent a diverse group of tumors with varying histological features and clinical behaviors. The assessment of these tumors demands a comprehensive understanding of anatomy, physiology, and proficiency in pathology due to their distinctive histological grading and clinical classifications(Choi et al., 2022). Adenoid cystic carcinoma is associated with delayed local recurrence, necessitating long-term patient follow-up. When dealing with a lesion in the palate, it is essential to consider adenoid cystic carcinoma in the differential diagnosis. The primary treatment goal is to achieve local control and prevent distant metastasis, underscoring the importance of early lesion detection for a favorable prognosis(Rafael et al., 2016).

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